Congenital Tracheal Occlusion in Neonates—A Minireview

KORAVANGATTU SANKARAN, MB, FRCP(C); CHANDER P. BHAGIRATH, MB; WALTER J. DUNCAN, MD; GORDON F. KASIAN, MD, *and* WILLIAM T. BINGHAM, MD, *Saskatoon, Saskatchewan*

Congenital tracheal occlusion, though seemingly rare, is generally fatal if not attended to immediately. A high index of suspicion should help to make the diagnosis in a neonate when there are respiratory distress at birth and no audible cry and an endotracheal tube cannot be advanced beyond the vocal cords. We present our experience with cases of congenital tracheal obstruction due to various causes during a period of four years.

(Sankaran K, Bhagirath CP, Duncan WJ, et al: Congenital tracheal occlusion in neonates—A minireview. West J Med 1985 Mar:142:348-351)

Congenital occlusion of the trachea is rare. If the problem is misdiagnosed, the invariable result is death of the newborn. Most of the infants previously described with congenital tracheal occlusions have survived for a few minutes to several days only. At the University Hospital, Saskatoon, during a period of four years ending December 1983, we have managed seven cases with congenital tracheal occlusions. Our objective in this paper is to review these cases and to deal with congenital tracheal occlusion as a separate disorder because in previous reports it was included with proximal upper airway occlusions, such as those of the oropharynx or larynx, and distal airway occlusions, such as bronchial and pulmonary agenesis.

Patients and Methods

Hospital records of infants admitted to the Neonatal Intensive Care Unit (NICU), University Hospital, Saskatoon, with the diagnosis of congenital tracheal occlusion were examined. During a period of four years ending December 31, 1983, a total of 2,920 infants were admitted. Of these infants, seven were identified as having congenital tracheal occlusion, presenting with immediate neonatal respiratory distress. The details of each case are given below. This tertiary-care center for sick neonates serves northern and central Saskatchewan, with 10,000 to 12,000 live births per year. The details of cases 1 and 2 have been described previously.[1,2]

Reports of Cases

CASE 1. Briefly, the patient, a baby boy, was born spontaneously at 36 weeks' gestation. The antenatal history included premature rupture of membranes and evidence of polyhydramnios. The infant was limp and cyanotic at birth. Attempts to intubate the trachea and oxygenate the child failed because the endotracheal tube would not pass beyond the vocal cords. Immediate tracheostomy improved the baby's condition dramatically. Later investigations confirmed the presence of proximal esophageal atresia, tracheal atresia just below the vocal cords and evidence of a tracheoesophageal fistula distally. The tracheoesophageal fistula was repaired on the first day of life. The atretic area of the trachea was resected at 1 month of age, and a stent was put in place. The stent had to be removed six weeks later, however, because of granulation tissue. At 2½ years of age, the infant is doing well with a tracheostomy. Bronchoscopy shows an opening of approximately 3 mm at the site of resection. Both physical and neurologic development of the baby are near normal for postconceptional age.

CASE 2. The patient, a female infant, was born by cesarean section 24 hours after the infant of case 1. The mother was at 36 weeks' gestation with a history of polyhydramnios. The infant at birth had no audible cry, was cyanotic and was in respiratory distress with pronounced indrawing. Attempts at intubation failed because the endotracheal tube would not pass beyond the vocal cords. An immediate tracheostomy resulted in remarkable improvement. As with case 1, investigations showed proximal esophageal and tracheal atresia with a distal tracheoesophageal fistula. The fistula was repaired immediately and, unlike the previous case, repair of the tracheal atresia was deferred to a later time. The infant is now 2½ years of age and doing remarkably well, her development

From the Neonatal Services, Department of Pediatrics, University of Saskatchewan, Saskatoon.

Presented in part at the Annual Meeting of the Royal College of Physicians and Surgeons of Canada, Quebec City, September 1982.

Submitted, revised, July 3, 1984.

Reprint requests to K. Sankaran, MB, Neonatal Services, Department of Pediatrics, University Hospital, Saskatoon, Saskatchewan S7N 0X0 Canada.

ABBREVIATIONS USED IN TEXT

CT = computed tomographic (scan)
NICU = Neonatal Intensive Care Unit

being normal, except for language skills, for her postconceptional age. Surgical correction is planned at about 4 to 5 years of age.

CASE 3. The patient, another term male infant, was born to a 21-year-old gravida 1, para 0 mother in a local hospital outside of Saskatoon. The mother received no antenatal care. Polyhydramnios was noted at the time of delivery. The baby was cyanotic despite pronounced respiratory effort and indrawing. Intubation was attempted twice, but again the endotracheal tube could not pass beyond the larynx. The infant died at 2 hours of age. Autopsy showed proximal esophageal atresia and a distal tracheoesophageal fistula with complete occlusion of the lumen of the larynx by a bar of cartilage immediately below the level of the vocal cords. A histologic section at the area of occlusion on the trachea showed a hairlike lumen lined with respiratory epithelium. No other congenital abnormalities were noted. Interestingly enough, this patient was born a month after the infants in cases 1 and 2 with the same abnormalities.

CASE 4. This female infant was born to a 24-year-old primigravid woman at 34 weeks' gestation in a breech presentation and of a breech delivery. An ultrasound study done at six months' gestation showed evidence of oligohydramnios and polycystic kidneys. The membranes ruptured at delivery and oligohydramnios was noted. Immediately after birth the baby showed respiratory distress, and attempts to pass an endotracheal tube failed. The Apgar scores were 5 at one minute and 0 at five minutes. An autopsy showed agenesis of the trachea along with agenesis of both lungs, the right kidney, ureter, urinary bladder and uterus. Other anomalies included a polycystic left kidney, an atrioseptal defect, polysplenia and a patent ductus arteriosus.

CASE 5. The patient, a male infant, was born to a 28-year-old gravida 2, para 1 mother at 32 weeks' gestation. An ultrasound study done antenatally showed oligohydramnios. The infant presented at birth with severe respiratory distress. Apgar scores were 1 at one minute and 0 at five minutes. An autopsy showed tracheal agenesis with agenesis of both lungs and polycystic kidneys.

CASE 6. This male infant was born to a 27-year-old gravida 2, para 2 mother by spontaneous vaginal delivery at term with no antenatal problems. The infant was cyanotic and grunting immediately after birth with a severe degree of indrawing and inspiratory wheeze. Apgar scores were 3 at one minute and 7 at five minutes. The infant was intubated with some difficulty by the transport team and taken to the NICU. A slight occlusion was noted in passing the endotracheal tube below the vocal cords. Later, extubation resulted in severe inspiratory stridor and cyanosis. An esophagogram, a computed tomographic (CT) scan of the neck and a chest x-ray film showed no abnormalities. Bronchoscopy showed narrowing of the lumen of the trachea below the vocal cords, suggesting an extrinsic compression and a severe degree of tracheomalacia at the site of the obstruction. Angiogram showed a partial vascular ring with an anomalous origin of the common carotid artery. Surgical suspension of the aortic arch was done in an attempt to relieve the extrinsic compression on the trachea. This produced no improvement. Tracheostomy was done to maintain ventilation. The plan is to remove the tracheostomy after six months following bronchoscopy. The infant is now 6 months old and there are no abnormalities on physical and neurologic examinations.

CASE 7. The patient, a male infant, was born at 36 weeks' gestation, weighing 1,650 grams, to a 24-year-old primigravid mother by cesarean section. Antenatal history was unremarkable except for breech presentation and a history of smoking, averaging half a pack a day through the pregnancy. Apgars were 4 at one minute and 8 at five minutes, and the baby required endotracheal intubation and ventilation. There was some difficulty noted in passing the tube beyond the vocal cords. The neonatal course was complicated by continual respiratory distress and evidence of congestive heart failure. Cardiac catheterization showed a patent ductus arteriosus, ventricular septal defect, dextrocardia and a prominent, high aortic arch causing tracheal compression. Bronchoscopy showed evidence of an extrinsic compression of the trachea. A surgical procedure was done to suspend the aortic arch and ligate the patent ductus arteriosus, with immediate improvement of respiratory state. Further investigations, such as bronchography, CT scan of the trachea and repeat bronchoscopy, showed significant stenosis of the entire trachea. Eventually, the baby was discharged at 10 months of age with minimal respiratory distress. He is normal physically and neurologically at 2½ years of age.

Discussion

The incidence of congenital tracheal occlusions at our center is about 1 in every 420 admissions. In other words, an approximate incidence of 1 in 5,000 to 7,000 live births is noted in the province of Saskatchewan. From our experience, congenital tracheal occlusion presents in three different forms: intraluminal—such as tracheal atresia, laryngeal web and failure to canalize the trachea—extraluminal due to external compression—that is, cystic hygroma, vascular rings, annular thyroid and the like—and tracheal maldevelopment, such as agenesis of the trachea and lungs.

Of the seven cases described above, three showed similar abnormalities. Two of these three babies recovered without sequelae. The third infant, even though he had a similar history, did not recover for two reasons. First, he was born at a small hospital where there were no facilities for immediate intervention and, second, the attending physicians were not sensitive to the problem. Because there was a hairlike lumen through the atretic area, the lungs were filled with air but there was inadequate gas exchange. The infant must have died of increasing acidosis, hypoxia and hypercapnia. The above cases have clearly shown that immediate intervention may be lifesaving. To our knowledge, these two infants are the first ever to have survived similar abnormalities beyond the neonatal period. Only one case with similar lesions has been reported before. This was diagnosed at autopsy.[3] The reason for clustering is still unknown. All investigations to elucidate this occurrence were unfruitful.

In the fourth and fifth cases, it was obvious that there was no palpable trachea, and this was associated with multiple congenital anomalies and oligohydramnios. Autopsy has confirmed anomalies inconsistent with sustained life.

The sixth case was one of aortic arch anomalies and associated tracheal occlusion and tracheomalacia at the site of occlusion. Again, the immediate neonatal presentation showed evidence of acute respiratory tract occlusion and difficult intubation. Tracheostomy was done to sustain the airway patency. The tracheomalacia due to compression is expected to resolve with time. Secondary tracheomalacia develops in those instances in which the tracheal wall has been subjected to chronic external pressure.[4]

The seventh case had tracheal stenosis and compression. As the infant grows, there will be relative abatement in the respiratory difficulty. This was confirmed by continued improvement observed in this case.

Congenital tracheal occlusion may occur when there are compressive extrinsic or intrinsic lesions of the trachea or tracheal maldevelopment. The extrinsic causes include cystic hygroma, goiter, tumors and vascular anomalies. Cystic hygroma is a cystic tumor of lymphatic tissue that frequently arises in the neck and may rapidly attain large size due to hemorrhage or infection. It may extend proximally into the floor of the mouth and distally into the mediastinum.[5] Congenital goiter is rare but usually seen in infants of mothers who have consumed large quantities of medication containing iodides during their pregnancy. In rare cases it can be large enough to cause respiratory obstruction at birth.[5]

Mediastinal cysts and tumors may produce acute respiratory occlusion at birth. Diagnosis is established radiologically, and the lesions may be differentiated by their position in the chest. Neurogenic tumors are usually in the posterior mediastinum—goiter and thymoma—and in the anterosuperior mediastinum; bronchogenic and esophageal duplication cysts are most frequently located in the hilar area, whereas the teratodermoid group of tumors is found in the anteroinferior mediastinum.[5]

The primary vascular malformations producing airway occlusion in infants are abnormalities of the aortic arch and its derivatives. Persistence of a vascular segment that would normally regress or abnormal regression of a portion that would normally remain patent may produce a vascular ring—this ring then causing occlusion of the trachea, esophagus or both.[6] The existence of symptoms depends, of course, on the relative tightness of the ring. The severity of the symptoms may vary from dysphagia to reflex apnea—so-called dying spells caused by tracheal occlusion.[7]

Of the four vascular lesions, three deserve discussion—double aortic arch, anomalous innominate artery and aberrant left pulmonary artery.

Of the patients with double aortic arch, 75% have symptoms varying from stridor with wheezing to dyspnea and cyanosis. Symptoms are made worse with crying and especially with feeding of solid foods. Diagnosis rests with angiography and the treatment is surgical.

The average age at surgical treatment is 8 months. About 95% of cases of double aortic arch involve division of the smaller persistent left aortic arch and left ligamentum arteriosum.[8]

A low-lying anomalous innominate artery can cause anterior tracheal compression. This group of patients suffers most from reflex apnea (16/23 in Toronto). Bronchoscopy will show the anterior tracheal compression as a pulsatile mass. The majority, 86% of these patients, have been treated conservatively[9] (Macdonald and Fearon, 1971) with gradual improvement. Persistence of reflex apnea requires surgical suspension of the innominate artery to the sternum, which provides nearly complete relief of symptoms.

If the pulmonary artery branch of the embryologic left sixth aortic arch fails to develop, a collateral branch from the right aortic arch then develops to supply the left lung, producing the very rare "aberrant" left pulmonary artery.[8] This vessel passes between the trachea and esophagus. The aberrant left pulmonary artery causes expiratory stridor in newborns as opposed to inspiratory stridor from the other types of rings. Diagnosis rests with angiography and simultaneous barium swallow. Treatment involves surgical repositioning of the aberrant left pulmonary artery.

The intrinsic causes of tracheal occlusion include subglottic hemangioma or, very rarely, lymphangioma and laryngeal webs. The diagnosis may be suggested by asymmetric narrowing of the trachea on chest x-ray film, but it should be confirmed by direct observation using endoscopy.[10] Case 7 exemplifies this point.

Tracheal anomalies such as stenosis, atresia and agenesis can present with respiratory tract occlusion at birth. Some of the lesions are amenable to immediate treatment, as evident in cases 1 and 2, whereas others are not. Atresia refers to congenital absence or closure of a normal opening, passage or cavity in the body, while agenesis refers to complete or almost complete failure of the anlage of an organ to develop. Severe cases of tracheal atresia often have been grouped with cases of tracheal agenesis.[11,12] We have classified cases 1 and 2 as tracheal atresia as we were unable to locate any opening on repeated bronchoscopies. Obviously, case 3 will be classified as severe stenosis or atresia, whereas case 7 has severe stenosis of the trachea. About 40 cases have been reported with tracheal atresia, agenesis or both in the literature.[12-15] Most had a tracheoesophageal or a bronchoesophageal fistula and, in several cases, surgical intervention was attempted.[15-18] All died eventually, with the longest survival being six weeks.[18]

The management of tracheal occlusion depends on awareness of the problem and prompt recognition. After it is established that an infant has acute respiratory tract occlusion and not respiratory depression, the airway should be cleared and a gentle attempt at ventilation should be tried. If intubation is unsuccessful, with difficulty encountered in advancing the endotracheal tube beyond the larynx, consideration should be given to immediate tracheostomy. The following features should alert attending physicians to immediate intervention: severe respiratory distress without audible cry, antenatal polyhydramnios or oligohydramnios, failure to advance an endotracheal tube beyond the vocal cords and palpable distal trachea. Once an airway is established, management will depend on the cause of the occlusion. Most cases will require surgical intervention at a later date.

In summary, physicians dealing with newborns must be familiar with the causes of acute tracheal occlusion in a neonate. Prompt diagnosis and proper management may help salvage some of these infants.

REFERENCES

1. Sankaran K, Bhagirath CP, Bingham WT, et al: Tracheal atresia, proximal esophageal atresia and distal tracheoesophageal fistula. Pediatrics 1983; 71:821-823

2. Sankaran K, Bhagirath CP, Bingham WT, et al: Tracheal atresia, proximal esophageal atresia and distal tracheoesophageal fistula: Two case reports. Presented at

the Royal College of Physicians and Surgeons of Canada Meeting in Quebec City, Canada, September, 1982

3. Ashley DJB: A case of congenital tracheal obstruction with esophageal atresia. J Pathol 1972; 108:261-263

4. Davies MRO, Cywes S: The flaccid trachea and tracheoesophageal congenital anomalies. J Pediatr Surg 1978; 13:363-367

5. Cywes S: Surgical respiratory emergencies in the newborn. S Afr Med J 1972; 46:1538-1544

6. Möes CAF: Anomalies of the aortic arch, *In* Keith JD, Rowe RD, Vlad P: Heart Disease in Infancy and Childhood. New York, Macmillan, 1978

7. Fearon B, Shortreed R: Tracheobronchial compression by congenital cardiovascular anomalies in children—Syndrome of apnea. Ann Otol Rhinol Laryngol 1963; 72:949-969

8. Berdon WE, Baker DH: Vascular anomalies and the infant lung: Rings, slings and other things. Semin Roentgenol 1972; 7:39-43

9. Macdonald RE, Fearon B: Innominate artery compression syndrome in children. Ann Otol Rhinol Laryngol 1971; 80:535-540

10. Kushner CA, Clifton-Harris GB: Obstructing lesions of the larynx and trachea in infants and children. Radiol Clin North Am 1978; 16:181-194

11. Floyd J, Campbell EC, Domini DE: Agenesis of the trachea. Am Rev Respir Dis 1962; 86:575-580

12. Faro RS, Goodwin CD, Organ CH, et al: Tracheal agenesis. Ann Thorac Surg 1979; 28:295-298

13. Warfel KA, Schulz DM: Agenesis of the trachea: Report of a case and review of the literature. Arch Pathol Lab Med 1976; 100:357-359

14. Hopkinson JM: Congenital absence of the trachea. J Pathol 1972; 107:63-67

15. Buchino JJ, Meagher DP, Cox JA: Tracheal agenesis: A clinical approach. J Pediatr Surg 1982; 17:132-137

16. Alfery DD, Ward CF, Plumer MH, et al: Management of tracheal atresia with tracheoesophageal fistula. Anesthesiology 1980; 53:242-244

17. Altman RP, Randolph JG, Shearin RB: Tracheal agenesis: Recognition and management. J Pediatr Surg 1972; 7:112-118

18. Fonkalsrud EW, Martelle RR, Maloney JV: Surgical treatment of tracheal agenesis. J Thorac Cardiovasc Surg 1963; 45:520-525